CASE REPORT

Juvenile female with chest wall fibromatosis located posteriorly to the right breast: radiation therapy or wait and watch?

Ali Akhavan,¹ Fariba Binesh,² Kargar Kargar,³ Hossein Navabii⁴

¹Department of Radiotherapy: Shahid Sadoghi University of Medical Sciences, Yazd, Iran

²Department of Pathology, Shahid Sadoghi University of Medical Sciences, Yazd, Iran

³Department of General Surgery, Shahid Sadoghi University of Medical Sciences, Yazd, Iran

⁴Shahid Sadoghi University, Yazd, Iran

Correspondence to

Dr Ali Akhavan,
ali52akhavan@yahoo.com

SUMMARY

Fibromatosis is a slow-growing monoclonal proliferation of fibroblasts. Although histologically benign it is locally invasive and can recur, however, without metastatic potential. Extra-abdominal fibromatosis is not common. Surgical resection is the cornerstone of treatment and radiation therapy is used in incomplete resection, however, its role is not proved through prospective randomised trials. This article presents a 17-year-old Iranian girl with chest wall fibromatosis located just behind the right breast. The patient was operated with positive margin, however, we decided to postpone radiation therapy because of potential risks.

take it seriously. Recently, the mass became larger and created numbness on her right arm when she bowed. Her medical history indicated no serious medical problem. There was no evidence of FAP in her family. Physical examination revealed a palpable soft mass originating from the right supraclavicular fossa extending to the chest behind the clavicle. Right arm movement and sensation was normal. CT scanning showed a well-circumscribed 60×80 mm mass in the supraclavicular fossa extending towards the apex of the chest just behind the right breast with no visible rib, lung and breast involvement (figures 1A, B).

BACKGROUND

Fibromatosis is a rare monoclonal proliferative disease of fibroblastic origin.¹ It is a slow-growing tumour with histologically benign appearance and without metastatic potential, however local invasion and recurrence is common.² It may be seen sporadically or in association with familial adenomatous polyposis (FAP). Incidence of sporadic type is 2–4 /million and 3.5–32% in FAP.³ Fibromatosis is classified into superficial (such as Dupuytren's, Ledderhose's and Peyronie's diseases) and deep also known as desmoids tumour.⁴ Desmoid tumours most commonly occur within the anterior abdominal wall in young women. Extra-abdominal sites are uncommon if occur, however they primarily involve muscles of the shoulder, pelvis and thigh.⁵ Chest wall is the origin of desmoids tumour in 10–28% of the patients.⁶ Surgery with wide local excision is the cornerstone of treatment, however, despite adequate margins, local recurrence is a significant problem that has been reported to range from 25% to 77% within the next 10 years postsurgery. The role of postsurgical irradiation in preventing relapse has been controversial.⁷ Despite the use of surgery and radiotherapy, 20–35% of patients with fibromatosis experience local recurrence. Therefore, different non-cytotoxic and cytotoxic drugs have been used with different efficacies.⁸ This report presents a 17-year-old girl with desmoids tumour of the chest wall just behind the breast and discusses controversies regarding the treatment.

CASE PRESENTATION

A 17-year-old Iranian girl referred to our centre owing to a soft tissue mass in the right supraclavicular fossa. The patient had noticed the mass since 1 year, however, because the mass was painless and did not cause any significant problem she did not

Figure 1 (A) Coronal view of tumour shows it locating in apex of the chest posterior to the breast. (White arrow shows the breast and black arrow shows the tumour.) (B) Axial view of tumour shows a well-circumscribed soft tissue mass locating anterior to the lung and posterior to the breast. (White arrows show the tumour and red marker shows the right breast.)

To cite: Akhavan A, Binesh F, Kargar K, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-006355

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of desmoids tumour includes reactive conditions such as scar and nodular fasciitis, and other mesenchymal neoplasms, such as low-grade fibrosarcoma and malignant nerve sheath tumour. Fortunately, nuclear staining for β -catenin greatly aids in the diagnosis and is a consistent finding (approximately 80% of cases), although it is not specific and is also seen in a rather large variety of other tumours. The diagnosis can be confirmed by screening for mutations (mainly in exon 3) of the β -catenin gene, which are found in about 85% of sporadic cases.

TREATMENT

Gross total resection was performed. The tumour was adhered to the neurovascular bundles without invading them. The tumour was shaved posteriorly and laterally from the anterior chest wall and the axillary fossa. Pathological examination revealed fibromatosis, however surgical margins were involved (figure 2). Owing to potential late effects of radiation, it was decided to postpone radiation therapy despite the existence of positive surgical margins and only sulindac 300 mg/day was started.

OUTCOME AND FOLLOW-UP

Now 3 months postsurgery the patient is well.

DISCUSSION

Desmoid tumours can be seen at any part of the body; however, the abdominal wall and soft tissues of the extremities, shoulder, neck and chest wall are sites of predilection.⁹ Although aetiology of these tumours is not exactly defined, genes, endocrinal secretions and trauma (mostly postoperative) might play a role.^{3 10} Intraabdominal desmoid tumours may present with intestinal obstruction and bowel ischaemia.¹⁰ Extra-abdominal tumours often present with abnormal firm masses and a pressure on contiguous structures³ as in this patient. CT scanning and MRI are useful in showing the relationship of the tumour to adjacent structures. Desmoid tumours have densities similar to muscle on contrast-enhanced CT scan; however, these tumours cannot be distinguished from other such similar soft tissue tumours. Although MRI is helpful in determining tumour extension and neurovascular involvement and is an accurate predictor of resectability, however it is of value for assessing the

Figure 2 Section shows interlacing bundles of fibroblasts separated by variable amounts of collagen.

tumour margins.³ In this patient CT scanning seemed enough for evaluation before surgery. The role of PET scanning has not been fully defined in the evaluation and management of this disease.¹⁰ Treatment decision for these patients is difficult because these tumours are rare and can involve various sites, their clinical course is variable and there is no prospective randomised trial to compare different treatment methods.^{1 3 9 10} A watchful waiting is a choice for asymptomatic tumours.¹⁰ Surgical excision remains the cornerstone of treatment, although resection with negative margin is the most accepted procedure,^{1 6 10} there are some controversies about the significance of microscopic positive margins.^{2 3 11–13} Adjuvant radiation therapy has been used in many studies owing to high probability of local recurrence despite negative surgical margins.^{1 3 7} On the other hand, most specialists propose radiation therapy when the tumour is not resectable, gross or microscopic when the tumour residue and recurrent disease are present.^{10 14 15} Although radiation therapy could be helpful, it has its own acute and late side effects. In this patient the mass was just posterior to the right breast and anterior to the right lung, the most important organs at risk of side effects especially at the late phase. Since the volume of the lung receiving high radiation dose is not significant and mean total lung dose is surely less than 20 Gy the lung toxicity is acceptable. Radiation therapy of the breast of young women, however is always of great concern. The most important issue is the risk of second cancers, carcinoma and sarcoma. Most of our knowledge about radiation-induced breast carcinoma in young girls revolves around studies on survivors of Hodgkin's lymphoma receiving radiation with various fields and doses with and without chemotherapy. In a study performed by the German Hodgkin's Lymphoma Study Group breast cancer was seen in 13 patients from 5367 treated patients. Sixty-two per cent of them were between 16 and 34 years at diagnosis/treatment of the Hodgkin's disease.¹⁶ In another study carried out by De Bruin *et al* among 1122 women, 5-year survivors treated for Hodgkin's disease before the age of 51, after a median follow-up of 17.8 years, 120 women developed breast cancer; indicating an absolute excess risk of 57/10 000 patients/year. The overall cumulative incidence 30 years after treatment was 19% (95% CI 16% to 23%); while for those treated before the age 21 years, it was 26% (95% CI 19% to 33%). The relative risk remained high after prolonged follow-up (>30 years posttreatment: standardised incidence ratio, 9.5; 95% CI 4.9 to 16.6). They concluded reduction in radiation volume appeared to decrease the risk of breast cancer after the Hodgkin's disease. Furthermore, women with ≥ 20 years of intact ovarian function after radiotherapy at young ages (<31 years) experienced significantly higher risks for breast cancer than those with fewer than 10 years of intact ovarian function.¹⁷ At a systematic review of the literature, girls and women with paediatric or young adult cancer (aged ≤ 30 years at diagnosis) who received chest radiation were assessed. From about 7000 women receiving chest radiation 422 (6%) women subsequently developed breast cancer. Increased risk for breast cancer was found as early as 8 years after chest radiation and did not plateau with increasing length of follow-up. The cumulative incidence of breast cancer by age 40–45 years ranged from 13% to 20% and by 25–30 years of follow-up ranged from 12% to 26%. This incidence is similar to that in women with a *BRCA* gene mutation, in whom by age 40, the cumulative incidence ranges from 10% to 19%.¹⁸ Although radiation-induced sarcomas subsequent to treatment of breast cancer are rare, they are of poor prognosis. According to a large-scale study performed at the Curie Institute (Paris, France),

among more than 16 700 patients with breast cancer who received breast radiation 35 cases of sarcomas were seen.¹⁹ Various drugs have been proposed for treating desmoid tumours with and without surgery and radiation therapy. Among them tamoxifen and sulindac are better known. Even different chemotherapy drugs have been used in unresectable and recurrent tumours with various degrees of effectiveness. In an Italian study 94 patients aged ≤ 21 years with desmoid tumours were evaluated. The patients were divided into three groups: with negative surgical margins, positive surgical margins and gross residue. Systemic therapy was given to 49 patients in the first or second line and the response rates were 47% and 50%, respectively.²⁰ Considering all parameters involved and after explanation of these facts to the patient and her family, we decided to limit treatment to only prescribing sulindac for the patient.

Learning points

- ▶ In deciding the treatment approach for the patients suffering from desmoids tumours, owing to controversies surrounding the significance of the positive surgical margins, the risk of aggressive surgery should be evaluated versus conservative surgery.
- ▶ Since most patients are young women the risk of secondary cancer subsequent to radiation therapy should be considered in such patients.
- ▶ However, rare desmoid tumour can be seen in the thoracic area and it presents a challenging step to determine its treatment approach.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Colombo C, Gronchi A. Desmoid-type fibromatosis: what works best? *Eur J Cancer* 2009;45:466–7.
- 2 Melis M, Zager JS, Sondak VK. Multimodality management of desmoid tumors: how important is a negative surgical margin? *J Surg Oncol* 2008;98:594–602.
- 3 Shields CJ, Winter DC, Kirwan WO, *et al.* Desmoid tumours. *Eur J Surg Oncol* 2001;27:701–6.
- 4 Chummun S, McLean NR, Abraham S, *et al.* Desmoid tumour of the breast. *J Plast Reconstr Aesthet Surg* 2010;63:339–45.
- 5 Schiller VL, Arndt RD, Brenner RJ. Aggressive fibromatosis of the chest associated with a silicone breast implant. *Chest* 1995;108:1466–8.
- 6 Yurtsever I, Topal U, Yalcin R, *et al.* Desmoid tumor of the chest wall. *Eur J Radiol* 2003;46:119–21.
- 7 Ozger H, Eralp L, Tokar B, *et al.* Evaluation of prognostic factors affecting recurrences and disease-free survival in extra-abdominal desmoid tumors. *Acta Orthop Traumatol Turc* 2007;41:291–4.
- 8 Janinis J, Patriki M, Vini L, *et al.* The pharmacological treatment of aggressive fibromatosis: a systematic review. *Ann Oncol* 2003;14:181–90.
- 9 Kasper B, Ströbel P, Hohenberger P. Desmoid tumors: clinical features and treatment options for advanced disease. *Oncologist* 2011;16:682–93.
- 10 Escobar C, Munker R, Thomas JO, *et al.* Update on desmoid tumors. *Ann Oncol* 2012;23:562–9.
- 11 Lev D, Kotilingam D, Wei C, *et al.* Optimizing treatment of desmoid tumors. *J Clin Oncol* 2007;25:1785–91.
- 12 Merchant NB, Lewis JJ, Woodruff JM, *et al.* Extremity and trunk desmoid tumors: a multifactorial analysis of outcome. *Cancer* 1999;86:2045–52.
- 13 Gronchi A, Casali PG, Mariani L, *et al.* Quality of surgery and outcome in extra-abdominal aggressive fibromatosis: a series of patients surgically treated at a single institution. *J Clin Oncol* 2003;21:1390–7.
- 14 Findikcioğlu A, Kılıç D, Koçer E, *et al.* Current therapies for extra-abdominal fibromatosis of the chest wall: a silent giant mass. *Turkish J Thorac Cardiovasc Surg* 2010;18:334–7.
- 15 Fontanesi J, Mott MP, Kraut MJ, *et al.* The role of postoperative irradiation in the treatment of locally recurrent incompletely resected extra-abdominal desmoid tumors. *Sarcoma* 2004;8:83–6.
- 16 Behringer K, Josting A, Schiller P, *et al.* German Hodgkin lymphoma study group. Solid tumors in patients treated for Hodgkin's disease: a report from the German hodgkin lymphoma study group. *Ann Oncol* 2004;15:1079–85.
- 17 De Bruin ML, Sparidans J, van't Veer MB, *et al.* Breast cancer risk in female survivors of Hodgkin's lymphoma: lower risk after smaller radiation volumes. *J Clin Oncol* 2009;27:4239–46.
- 18 Henderson TO, Amsterdam A, Bhatia S, *et al.* Systematic review: surveillance for breast cancer in women treated with chest radiation for childhood, adolescent, or young adult cancer. *Ann Intern Med* 2010;152:444–55.
- 19 Kirova YM, Vilcoq JR, Asselain B, *et al.* Radiation-induced sarcomas after radiotherapy for breast carcinoma: a large-scale single-institution review. *Cancer* 2005;104:856–63.
- 20 Meazza C, Bisogno G, Gronchi A, *et al.* Aggressive fibromatosis in children and adolescents: the Italian experience. *Cancer* 2010;116:233–40.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow